Eisenmenger Syndrome in Pregnancy

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Summary

In last 10 years, only 3 cases of Eisenmenger syndrome in pregnancy were found. All of them were treated with conventional therapy and all 3 delivered vaginally. Two of the 3 babies were premature. Two of the 3 mothers and all 3 babies survived. The only maternal death was postpartum.

Introduction

In 1958, Wood (1958) expanded the anatomic concept of the Eisenmenger complex and defined Eisenmenger syndrome (ES) as "pulmonary hypertension at systemic level, due to a high pulmonary vascular resistance with reversed or bidirectional shunts at aorto-pulmonary, ventricular or atrial level".

This syndrome in pregnancy and especially in puerperium carries a high maternal and fetal morbidity and mortality (Gleisher et al 1979). Maternal mortality in cases with Eisenmenger's syndrome has changed very little over last 50 years (Yentis et al, 1998). The mainstay of treatment is administration of oxygen, digitalis, diuretics and antibiotics (conventional therapy). The use of heparin and sympathomimetics is still controversial. Administration of nitric oxide by inhalation during labor lowers pulmonary artery pressures within minutes and improves oxygenation (Goodwin et al 1999). We came across 3 cases in last 10 years who were treated with conventional therapy, 2 of the mothers and all the 3 babies survived.

Case reports

Case No. 1

Mrs A, 20 years of age, a primigravida presented at 20 weeks of gestation with palpitation and progressive effort dyspnoea. She had cyanosis of the digits and tongue since childhood. On examination she had clubbing of fingers and toes and central cyanosis. There was clinical evidence of right ventricular hypertrophy and pulmonary hypertension. Obstetrical examination revealed a gravid uterus corresponding to gestational age. The electrocardiogram and chest X-ray were consistent with features of ES. A 2 dimensional echocardiogram demonstrated ventricular septal defect with pulmonary hypertension. Haemogram, blood biochemistry and urine analysis were within normal limits. Serial ultrasonography and nonstress test were

done to assess fetal well being. Patient was given digitalis, Lasix, KCl, antibiotics and oxygen. Labour was induced at term and a mature female infant was delivered by vacuum extraction. Throughout labor, haemodynamics remained stable. Her cyanosis became worse in early puerperium but she responded well to conventional therapy and was discharged.

Case No. 2

Mrs. B, a 3rd gravida 23 years of age, presented at 32 weeks of gestation with progressive effort dyspnoea. She was known to have had heart disease since early childhood manifested by cyanosis on exercise and by frequent respiratory infections. She had two preterm deliveries at home at 30 weeks of gestation. Both the babies died within a week of birth. On examination she had central cyanosis, clubbing of fingers, loud pulmonary component of second heart sound on auscultation and right ventricular hypertrophy as documented by chest X-ray and electrocardiography. A 2-dimensional echocardiography showed large atrial septal defect with bidirectional shunt. Ultrasonography and nonstress test were done to assess fetal well being. She went into preterm labor at 34 weeks gestation. Tocolysis was started and dexamethasone given. Two days later she delivered vaginally a premature female baby. Patient's condition worsened twice during puerperium. She was managed by conventional therapy to which she responded well and was discharged after 2 weeks.

Case No. 3

Mrs C, primigravida, 24 years of age apparently asymptomatic and unaware of any cardiac disease till 10 days back when she went to a hospital at 32 weeks of gestation with bluish discoloration of nails, palpitation and breathlessness. She was diagnosed there to be a case of ES and referred to this hospital. She presented with preterm labor and liquor draining per vagina for 10 hours. On examination, she had clubbing of fingers and toes with central cyanosis and BP was 140/96. She was febrile. There was clinical evidence of right ventricular hypertrophy and pulmonary hypertension. Obstetrical examination showed a gravid uterus corresponding to 28 weeks of gestation. She was in labour with absent membrane. Electrocardiography and chest X-ray were suggestive of ES. A 2-dimensional echocardiography showed ventricular septal defect. Labour was augmented with syntocinon. She delivered vaginally a premature male baby. She was treated with antibiotics, digitalis, oxygen, frusemide and nifedipine. Patient developed hypotension on 3rd postnatal day and was shifted to intensive care unit for management. She recovered from this episode. On 16th postnatal day again her condition deteriorated and she succumbed to death. Autopsy was not performed as patient's relatives refused to give consent.

Discussion

The symptomatology of ES though partially depends upon the underlying cardiac defect, the common finding of pulmonary hypertension will be there in all cases. Lack of symptoms may be striking and the condition may not be diagnosed until pregnancy as in case No. 3. This feature was also noted by Gleicher et al (1979). The diagnosis in all our patients was made by the clinical findings and supported by ECG, 2-D echocardiograph and X-ray chest.

The most accepted explanation for high maternal mortality especially in the early postpartum period, is hypotension which leads to increase in right to left shunt following delivery (Jones & Howitt 1965, Pitts et al 1977). It is obvious that factors that increase pulmonary resistance, decreases systemic resistance or causes a fall in peripheral blood pressure through volume loss which enhances the right to left shunt. Such a change may occur acutely, as with a sudden post partum blood loss or pulmonary embolus, or may be caused by the inability of the pulmonary vasculature to readjust to pregnancy hemodynamic condition. Pre eclampsia is known to aggravate the hypercoagulable state of pregnancy even more. The only case of ours which died during puerperium suffered from pre eclampsia. As previously reported by others (Jones & Howitt 1965, Arias 1977, Crawford et al 1971) maternal mortality was highest during parturition and in the puerperium; sudden death has been described upto 22 days after delivery (Neilson et al 1971).

Jeyamalar et al (1992) concluded that the patients who were managed by conventional therapy succumbed to death and those managed by careful use of pharmacological agents namely aramine and heparin survived. However, our experience has proved that conventional therapy is quite effective. The use of anticoagulation in fact is sharply opposed by Nanayakkara and Pieris (1964) and later by Pitts et al (1977). Lust et al (1999) and Goodwin et al (1999) have shown that their patients responded well to inhalation of nitric oxide, but in both cases mother died in puerperium. We have not used anticoagulant or nitric oxide in our cases. Calcium antagonists are known to decrease pulmonary arterial pressure more than systemic arterial pressure in ES (Wong et al 1989) so we used nifedipine in our 3rd case which had PIH.

The normal pregnant patient tolerates the

sudden blood loss of upto 1 litre without much problem but a patient of ES may not be able to adjust her pulmonary circulation. This will increase her right to left shunt, decreased cardiac output and she will enter a vicious cycle. This is the hypothetical explanation for high maternal mortality in association with caesarean section (Gleisher et al, 1979, Jones & Howitt 1965). Hence, vaginal delivery seems to be the mode of choice. All our 3 cases delivered vaginally. Gleisher et al (1979) in his review has shown that more than half of all deliveries were premature. Whereas Yentis et al (1998) have shown that only 15% of infants were born at term. The total perinatal mortality reached 28.3% which was mainly because of prematurity. Two of our 3 cases had preterm labour, however the babies survived as they were not too premature. Since sudden death is common in early puerperium it is advisable to keep the patients in hospital for 2-3 weeks after delivery.

In a case of ES every obstetrician should counsel the patient against pregnancy. It is so important that Gleisher et al (1979), have given provocative statement like "abortion is the proper management for pregnancy" which is very true. A first trimester D & C under paracervical anaesthesia should be performed for interruption of pregnancy. Elective surgical sterilization, no less than three months after any previous pregnancy is advisable. Local and conduction anaesthesia are preferable to general anesthesia (Arias, 1977). Both our patients who survived were counseled against further pregnancy were advised to come back after 3 months. Due to lack of good antenatal care all the 3 reported cases were diagnosed late in pregnancy. Two of them responded to conventional therapy. These patients should normally be advised against pregnancy. If they come with pregnancy, abortion in I trimester or elective sterilization after delivery should be done.

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